occasionally be difficult. Two general rules of thumb are helpful:

- Benign tumors usually produce obvious vascular channels filled with blood cells (e.g. capillaries filled with red cells) lined by a monolayer of normal-appearing endothelial cells.
- Malignant tumors are more cellular and more proliferative, and exhibit cytologic atypia; they usually do not form well-organized vessels. The endothelial derivation of neoplastic proliferations that do not form distinct vascular lumina can usually be confirmed by immunohistochemical demonstration of endothelial cell-specific markers such as CD31 or von Willebrand factor.

Benign Tumors and Tumor-Like Conditions

Vascular Ectasias. *Ectasia* is a generic term for any local dilation of a structure, while *telangiectasia* is used to describe a permanent dilation of preexisting small vessels (capillaries, venules, and arterioles) that form a discrete red lesion—usually in the skin or mucous membranes. These can be congenital or acquired and *are not true neoplasms;* some of them are malformations and others are hamartomas:

- *Nevus flammeus* (a "birthmark"), the most common form of vascular ectasia, is a light pink to deep purple flat lesion on the head or neck composed of dilated vessels. Most ultimately regress spontaneously.
- The so-called *port wine stain* is a special form of nevus flammeus. These lesions tend to grow during childhood, thicken the skin surface, and do not fade with time. Such lesions in the distribution of the trigeminal nerve are associated with the *Sturge-Weber syndrome* (also called *encephalotrigeminal angiomatosis*). This uncommon congenital disorder is associated with facial port wine nevi, ipsilateral venous angiomas in the cortical leptomeninges, mental retardation, seizures, hemiplegia, and skull radio-opacities. Thus, a large facial telangiectasia in a child with mental deficiency may indicate the presence of additional vascular malformations.
- *Spider telangiectasias* are nonneoplastic vascular lesions grossly resembling a spider. These manifest as radial, often pulsatile arrays of dilated subcutaneous arteries or arterioles (resembling spider legs) about a central core (resembling a spider's body) that blanch with pressure. These commonly occur on the face, neck, or upper chest and are most frequently associated with hyperestrogenic states, such as pregnancy or liver cirrhosis.
- *Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu disease)* is an autosomal dominant disorder caused by mutations in genes that encode components of the TGF-β signaling pathway. The telangiectasias are malformations composed of dilated capillaries and veins that are present at birth. They are widely distributed over the skin and oral mucous membranes, as well as in the respiratory, gastrointestinal, and urinary tracts. The lesions can spontaneously rupture, causing serious epistaxis (nosebleed), gastrointestinal bleeding, or hematuria.

Hemangioma. Hemangiomas are very common tumors characterized by increased numbers of normal or abnormal vessels filled with blood (Fig. 11-30). These lesions constitute 7% of all benign tumors of infancy and childhood; most are present from birth and initially increase in size, but many eventually regress spontaneously. While hemangiomas typically are localized lesions confined to the head and neck, they can occasionally be more extensive (*angiomatosis*) and can occur internally. Nearly one third of these internal lesions are found in the liver. Malignant transformation is rare. Several histologic and clinical variants have been described:

- *Capillary hemangiomas* are the most common type; these occur in the skin, subcutaneous tissues, and mucous membranes of the oral cavities and lips, as well as in the liver, spleen, and kidneys (Fig. 11-30*A*). Histologically, they are composed of thin-walled capillaries with scant stroma (Fig. 11-30*B*).
- *Juvenile hemangiomas* (so-called "strawberry type" hemangiomas) of the newborn are extremely common (1 in 200 births) and can be multiple. These arise in the skin and grow rapidly for a few months, but then fade by 1 to 3 years of age and completely regress by age 7 in the vast majority of cases.
- *Cavernous hemangiomas* are composed of large, dilated vascular channels. As compared to capillary hemangiomas, *cavernous hemangiomas* are more infiltrative, frequently involve deep structures, and do not spontaneously regress. On histologic examination, the mass is unencapsulated, has infiltrative borders, and is composed of *large, cavernous blood-filled vascular* spaces separated by connective tissue stroma (Fig. 11-30*C*). Intravascular thrombosis and associated dystrophic calcification are common. They can be locally destructive, and as a result some may require surgery. More often the tumors are of little clinical significance, but they can be cosmetically troublesome and are vulnerable to traumatic ulceration and bleeding. Moreover, cavernous hemangiomas detected by imaging studies may be difficult to distinguish from their malignant counterparts. Brain hemangiomas are also problematic, as they can cause symptoms related to compression of adjacent tissue or rupture. Cavernous hemangiomas are one component of *von Hippel-Lindau disease* (Chapter 28), in which vascular lesions are commonly found in the cerebellum, brain stem, retina, pancreas, and liver.
- *Pyogenic granulomas* are capillary hemangiomas that present as rapidly growing red pedunculated lesions on the skin, gingival, or oral mucosa. They bleed easily and are often ulcerated (Fig. 11-30*D*). Roughly a quarter of lesions develop after trauma, reaching a size of 1 to 2 cm within a few weeks. Curettage and cautery is usually curative. *Pregnancy tumor* (granuloma gravidarum) is a pyogenic granuloma that occurs infrequently (1% of patients) in the gingiva of pregnant women. These lesions may spontaneously regress (especially after pregnancy), or undergo fibrosis, but occasionally require surgical excision.

Lymphangiomas. Lymphangiomas are the benign lymphatic counterparts of hemangiomas.

- *Simple (capillary) lymphangiomas* are slightly elevated or sometimes pedunculated lesions up to 1 to 2 cm in diameter that occur predominantly in the head,